Title: Temporal Encephalocele: A treatable etiology of pharmacoresistant pediatric temporal lobe epilepsy

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***Purpose*:** Temporal lobe encephaloceles (TEs) are defined as a pathological herniation of brain parenchyma through a structural defect in the middle cranial fossa. They are a rare cause of pharmacoresistant temporal lobe epilepsy in adults but not well described in pediatric epilepsy. The purpose of this study is to describe temporal encephalocele in children with temporal lobe epilepsy, to outline radiographic features of these cases, and to report surgical treatments in our cohort.
***Materials and Methods*:** We retrospectively reviewed patients with epilepsy and temporal encephalocele on brain imaging, who were between 0 and 18 years of age at Mayo Clinic Children's Center from 2008 to 2018. Data was collected on demographics, brain imaging features and surgical outcomes.
Results: Eight patients diagnosed with temporal encephaloceles (TE) were identified. The median age at epilepsy onset was 10.5 years [IQR 8.5-13.5 years]. Latency between epilepsy diagnosis and definite TE diagnosis had a median of 4 years (range of 0-17 years). Our patient series had a mean BMI of 26. Half of the patients had risk factors for seizures including fever, family history of seizures, previous traumatic brain injury, or perinatal infection. The most common seizure types were focal epilepsy with or without secondary generalization with temporal region onset. Our entire cohort had associated comorbidities including depression, anxiety and learning disabilities. Of the patients who had surgery (6/8), 83% (5/6) were seizure free with Engel outcome IA with mean duration of 16.7 months follow-up after surgery. One patient had more than 90% seizure frequency reduction up to 2 years after encephalocele resection and repair. More than one third (38%) of the patients were diagnosed based on epilepsy surgery conference re-review. Three quarters of the patients were on an average of two antiepileptic drugs at the time of diagnosis. All of the patients (8/8) had protrusion of CSF and distorted/gliotic brain parenchyma through an osseous defect in the floor of the middle fossa without frank osseous dehiscence. In 50% of the cases, there was associated regional FDG hypometabolism on brain PET imaging. No patient had bilateral TE.
Conclusions: We conclude that temporal lobe encephalocele is a missed cause of drug-resistant temporal lobe epilepsy in childhood. All patients had a unilateral temporal lobe encephalocele. All herniations were contained defects without osseous dehiscence. Of the patients who had surgery, 83% were seizure free at last follow-up.